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lesson of the month (2)

Syncope resistant to anticonvulsant therapy

It is recognised that physicians can face difficulty in identifying the aetiology of an episode of loss of consciousness. This lesson reports the case of a patient who was diagnosed with epilepsy in 2008 after presenting with 'funny turns' and had more than 100 further blackouts before a correct diagnosis of sinus node disease was made in 2010. Implantation of a permanent pacemaker has abolished these episodes and restored the patient's quality of life. Diagnostic and management strategies are discussed for patients presenting with loss of consciousness. Physicians are reminded of the difficulty in distinguishing cardiovascular syncope from epilepsy in such patients and the broader principle of questioning a diagnosis in those who do not appear to respond to treatment.

Lesson

A 71-year-old retired teacher was referred for cardiac assessment in 2008 having suffered two episodes of transient loss of consciousness (T-LOC). Both episodes had occurred without warning and he denied preceding chest pain, palpitations or light headedness. One had occurred while standing, the other while sitting. His wife had witnessed both and reported twitching of his limbs while unconscious and disorientation on first regaining consciousness. There was no tongue biting, incontinence or prolonged drowsiness. He had a past history of inferior ST-segment elevation myocardial infarction, hypertension and bipolar disorder. At the time his medications were aspirin 75 mg daily, simvastatin 40 mg daily, ramipril 10 mg daily, bisoprolol 2.5 mg daily and lithium.

His resting 12-lead electrocardiogram (ECG) was within normal limits. Blood tests revealed normal thyroid function and normal serum lithium levels. The patient was asymptomatic during a 48-hour ambulatory ECG, which revealed several paroxysms of atrial fibrillation (AF) but no prolonged pauses or bradyarrhythmias. Carotid duplex scanning was unremarkable.

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In the absence of a readily identifiable cardiovascular aetiology, he was referred on to a neurologist. It was thought that the episodes were most likely to represent complex partial seizures and thus the patient started antiepileptic medication (lamotrigine). The episodes of T-LOC continued, however, and so the dose of lamotrigine was increased and subsequently changed to levetiracetam. Two electroencephalograms (EEG) performed six months apart showed no evidence of epileptiform activity. By May 2010, over two years after first presenting, these episodes were still occurring and the diagnosis of epilepsy was questioned.

The patient was thus re-referred to cardiology and, given that his symptom frequency had increased markedly over the past two years, a repeat 48-hour Holter monitor was requested. This revealed frequent paroxysms of AF with numerous pauses on cardioverting from AF to sinus rhythm, the longest of which was a 24-second period of asystole (Fig 1). The patient was immediately contacted by telephone, urgently admitted and a permanent pacemaker was implanted the following day for treatment of his sinus node disease. At one-month follow-up, the patient has had no further collapses. The patient estimates that he suffered over 100 such blackouts in two years, including five in the two days prior to his admission. Fortuitously, he never sustained a serious injury.

Discussion

T-LOC is common – around 40% of adults have experienced an episode of syncope, and syncope accounts for 6% of emergency medical admissions.[1] Cardiovascular syncope is far more common than epilepsy.[2] Myoclonic jerks are common during cerebral hypoperfusion irrespective of aetiology and are not helpful in discriminating epilepsy from other causes of syncope.[3] This patient had sinus node dysfunction (SND) and its

Key learning points

- Cardiovascular syncope is more common than epilepsy as a cause of transient loss of consciousness (T-LOC).
- Myoclonic jerks are common in any case of cerebral hypoxia and should not be used to exclude cardiac syncope.
- Ambulatory electrocardiogram (ECG) monitoring must be correlated with a patient symptom diary: absence of symptoms during a brief monitored period does not mean absence of conduction system disease.
- Consider implantable loop recorders in cases of infrequent symptoms.
- Always reconsider a diagnosis if initial management strategies are failing.

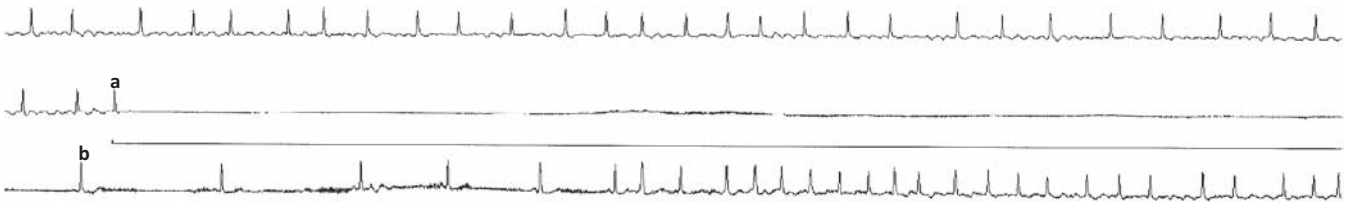

Fig 1. Extract from patient's 48-hour ambulatory electrocardiogram monitor revealing start (point a) and end (point b) of a 24-second period of asystole. Note atrial fibrillation (AF) before the pause followed by a few sinus beats and then re-initiation of AF (irregularly irregular QRS complexes).

most dramatic presentation, namely syncope. It is known that the majority of patients with syncope due to SND will experience recurrent syncope[4] and the only effective treatment in this situation is permanent cardiac pacing.[5]

Diagnostic difficulty in distinguishing epilepsy from cardiovascular syncope is well recognised. One British study found that 42% of patients previously diagnosed with epilepsy actually had a cardiovascular abnormality leading to T-LOC.[6] A report from the All Party Parliamentary Group on epilepsy in 2007 described the estimated British misdiagnosis rate of 23% as 'shocking'.[7] This is an important issue for patients as the diagnosis of epilepsy has associated implications for driving, insurance, social stigma and possibly occupation.[8] Obtaining a detailed history, with bystander witnesses if possible, is crucial.

All patients with T-LOC should have a 12-lead ECG and, if abnormal, an echocardiogram. Cardiovascular investigations which may be indicated include head-up tilt table testing and implantable loop recorder (ILR) insertion. ILRs offer the advantage of compliance and convenience during long-term monitoring efforts as symptoms can be very sporadic.[9] Routine ambulatory ECG monitoring in all T-LOC patients has a very low yield[10] and a normal result should not be used to exclude conduction abnormalities if the patient was asymptomatic during the recording. The 2009 European Society of Cardiology syncope guidelines only recommend ambulatory ECG monitoring if symptoms are occurring very frequently (defined as one or more episodes weekly).[11] Additionally, the latest guidelines from the National Institute for Health and Clinical Excellence (NICE) do recommend ILR insertion for episodes occurring less than once fortnightly[12] – this would have been the case with this patient at the time of his first presentation and would, undoubtedly, have lead to a more prompt diagnosis. In those patients in whom symptomatic bradycardia is detected, appropriate pacing both improves quality of life[13] and reduces mortality in those with documented atrioventricular block without a reversible cause.[14] It is clear that delays in putting patients forward for pacing are commonplace[15] and may result in harm.[16]

In this case, the decision to pace was made two years after the initial episode of syncope. This was largely due to an alternative diagnosis being reached. The impact this had on the patient and his wife cannot be overstated. They feel their 'world was turned upside-down'. They avoided public places, such as theatres and cinemas, in case he had an attack and, when outdoors, he wore a hat lined with foam to avoid serious head injury in case he fell to the ground. The patient's wife feels that their lives have returned to normal again following correct diagnosis and treatment.

Conclusion

Reaching the correct diagnosis in patients with T-LOC is often difficult, but incorrect diagnosis results in delays to appropriate management with potential for harm caused either by ongoing symptoms or the risks of unnecessary treatment. This case highlights the importance of symptom-rhythm correlation on ECG before excluding rhythm disturbance as the cause of syncope, but it also reminds us of the broader medical principle of questioning a diagnosis in patients who fail to respond to initial therapy.

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book reviews

Sherlock's disease of the liver and biliary systems, 12th edn
Edited by James S Dooley, Anna Lok, Andrew Burroughs, Jenny Heathcote. Wiley Blackwell, Oxford 2011. 792 pp. £130.00.

One naturally mourns for the singlehanded authority of the late Sheila Sherlock in her famous textbook. But one has to accept that the extraordinary growth of hepatology since then demanded a greater pool of contributing expertise. James Dooley, who co-authored the last three editions of the textbook with Sheila, has brought in three additional editors for this new 12th edition – Anna Lok from the USA, Andrew Burroughs from the UK and Jenny Heathcote from Canada. They have overseen a large team of outstanding experts covering the whole specialty as it is today. Many of the contributors were trained or had worked with Sheila at different times and coming, as it were, from the same mould have maintained that didactic approach of presentation that brought the book such success over its 50 years of existence. 'An accurate source to relevant information from students to specialist physicians' is how it is described in the preface of this new volume.

The clarity of figures and tables has always been a hallmark of the book and the production team at Wiley-Blackwell is to be congratulated on the standards obtained. The superb artwork evident in the first chapter by J Lefkowitch on anatomy and function is as exciting as is the text. In fact, whichever chapter one dips into one wants to continue reading. It is invidious to isolate chapters. One on hepatic fibrogenesis by Scott Freidman, for example, makes light of scientific complexities and is truly informative. Those by Andrew Burroughs on portal hypertension, Marsha Morgan's on encephalopathy, Jenny Heathcote's on autoimmune disease and Anna Lok's on hepatitis B, and the editors' own contributions, are outstanding authoritative accounts, reflecting the vast clinical experience, as well as research, contributions in these areas.

There are useful contributions on special topics including the liver and systemic disease, and the liver in pregnancy which will prove useful to clinicians facing individual clinical problems. The hepatic transplantation chapter has been extended, reflecting the importance of this form of therapy in current hepatological practice.

James Dooley is to be congratulated on his courage and dedication along with his clearly inspiring leadership to contributing authors, and he has not allowed Sheila's volume to die with her passing. The hepatology world has been given a thoroughly up-to-date, exciting and cleverly produced volume which competitors will have the greatest difficulty in matching.

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